Radiology Corner (Case #4)

Left Atrial Myxoma

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Introduction

Cardiac myxomas are benign tumors of the heart and arise most commonly from the interatrial septum, typically on the left side. They may grow to be fairly large at presentation, often associated with intracardiac obstruction, embolic events, or constitutional symptoms. Although they are histologically benign, cardiac myxomas by virtue of their location and size can be a source of significant morbidity and mortality. Notably, patients with cardiac myxomas have an increased risk for sudden death and surgical excision is typically recommended.

Primary tumors of the heart are rare, but are typically benign when they do occur. Approximately half of benign primary tumors of the heart are cardiac myxomas. Cardiac myxomas may remain clinically occult for many years, resulting in their substantial growth prior to manifesting symptoms. Their large size at presentation and intracavitary location can be associated with significant morbidity and mortality, even sudden death. Cardiac myxomas, therefore, require urgent attention and surgical excision.

History

A 51-year-old woman was referred from a satellite medical clinic for the evaluation of dyspnea on exertion. Her symptoms had significantly worsened since her travel abroad 4 weeks ago and now included shortness of breath even with conversation. The patient also reported experiencing three upper respiratory infections in the past 4 months each requiring medical treatment, the last episode just 1 week prior to this admission with a fever to 102 ˚F. The patient had otherwise no other significant past medical history. Physical exam was notable for a temperature of 97 ˚F and a faint 2/6 holosystolic heart murmur. The patient was admitted for further evaluation of her worsened respiratory status. PA (Figure 1A) and lateral (Figure 1B) chest radiographs were obtained and the patient also underwent a contrast-enhanced pulmonary CT angiogram (CTA; Figure 1C, lung windows; Figure 1D, soft tissue windows) for evaluation of possible pulmonary embolism.

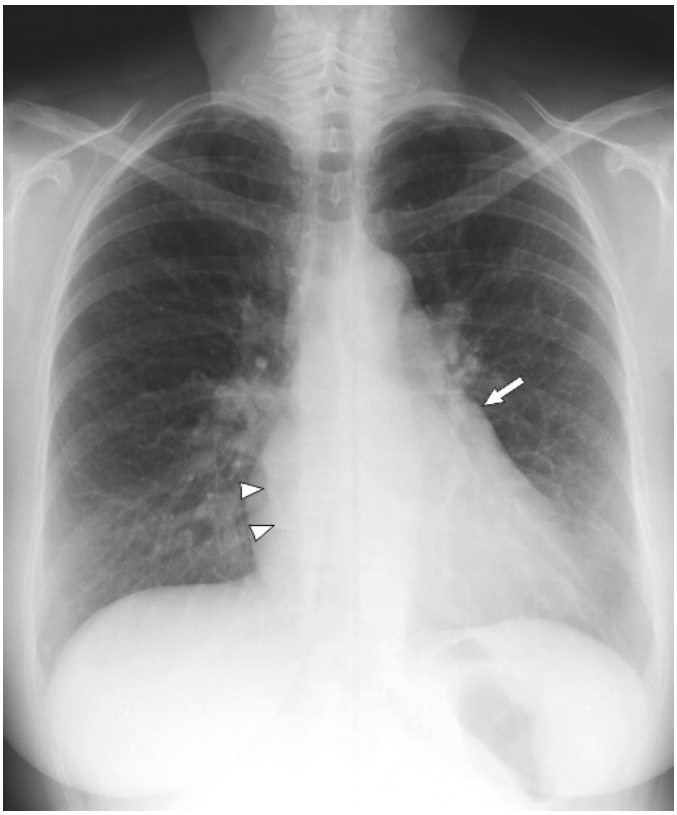

Figure 1A

Figures 1A. Chest radiographs demonstrated signs of left atrial enlargement. On the PA chest radiograph, a double density (arrowheads) consistent with an enlarged left atrium was noted superimposed over the cardiac silhouette. The double density is most evident over the right heart shadow. Enlargement of the left atrial appendage was also noted (white arrow).

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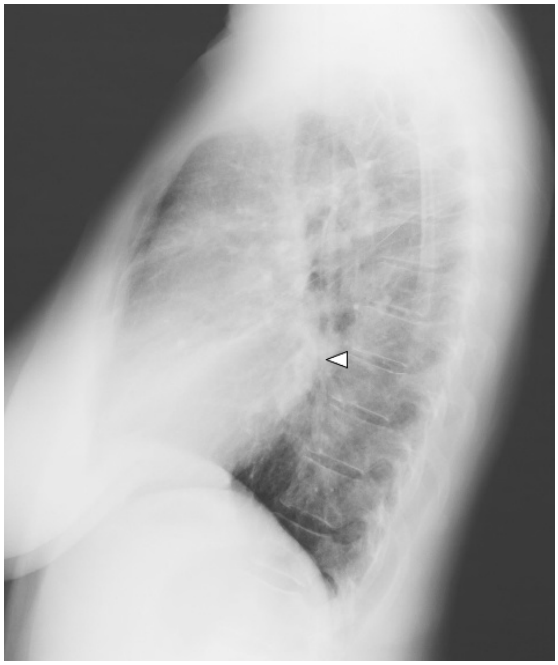

Figure 1B

Figures 1B. On the lateral chest radiograph, there is subtle posterior bulging of the cardiac silhouette (arrowhead), which suggests left atrial enlargement.

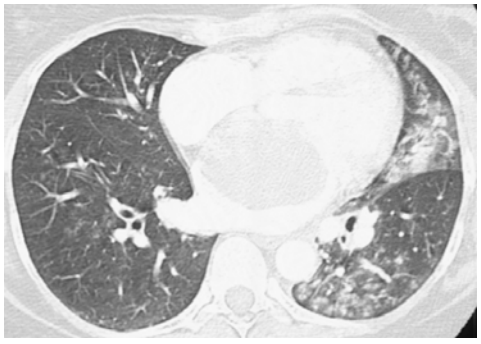

Figure 1C

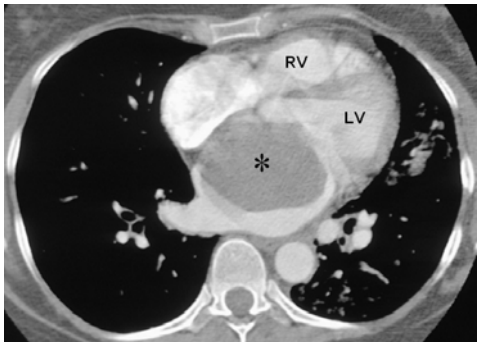

Figure 1D

Figures 1C and 1D. Axial image from a contrast-enhanced pulmonary CT angiogram demonstrated normal pulmonary arteries (i.e. no pulmonary embolus) and confirmed the presence of left-sided pulmonary airspace disease (1C, lung windows). On soft tissue windows (1D), the large 5.5 x 6.5 cm ovoid mass (*) was noted in the left atrium adjacent to the interatrial septum. Incidentally, the patchy left upper and lower lobe infiltrates resolved fairly quickly following surgery (not shown) and were therefore thought to primarily represent pulmonary congestion from obstruction of the left atrium by the large myxoma. RV = right ventricle; LV = left ventricle

Imaging Findings

The PA chest radiograph (Figure 1A) demonstrates an abnormal density overlying the right heart shadow (double-density sign) and a prominent left atrial appendage, suggestive of left atrial enlargement. Contrast-enhanced pulmonary CTA demonstrated normal pulmonary arteries without evidence for pulmonary embolism but note of patchy airspace disease within the left upper and lower lobes (Figure 1C), which represented areas of patchy pulmonary edema secondary to elevated left atrial pressures. A large 5.6 cm x 6.5 cm ovoid mass with soft tissue density (25 Hounsfield units) was noted in the left atrium on soft tissue windows (Figure 1D). The mass was well circumscribed but lobular in contour and abutted the inter-atrial septum.

An MRI study was also performed. On cine bright blood gradient echo images (Figures 1E and 1F), the large left atrial mass had a homogeneous dark signal intensity and was noted to be attached to the interatrial septum. The mass obstructed the mitral valve but frank movement of the mass across the valve was not seen. Compared to myocardium, the mass was relatively isointense but heterogeneous in its signal intensity on T1-weighted black blood images (1G). Following the intravenous administration of 0.1 mmol/kg dose of gadolinium-chelate contrast media, the mass enhanced heterogeneously (Figure 1H). The patient underwent successful removal of the mass and the diagnosis of cardiac myxoma was confirmed pathologically.

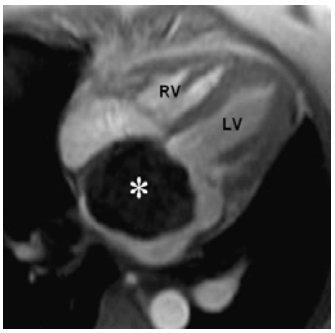

Figure 1E

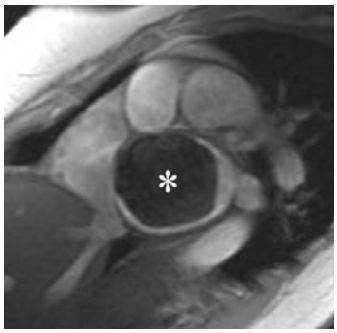

Figure 1F

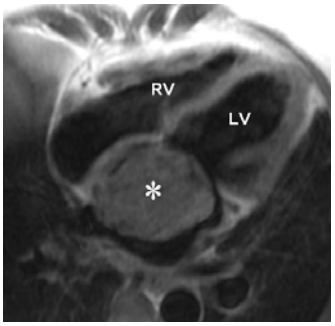

Figure 1G

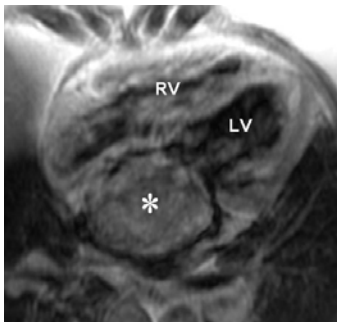

Figure 1H

Figures 1E to 1H. On cardiac-gated, cine gradient echo "bright blood" MR images (1E, long axis view; 1F, short axis view), a large hypointense mass (*) is noted in the left atrium. The mass was not very mobile and did not have a long stalk. On cine imaging, the mass did not cross the mitral valve and did not prolapse into the left ventricle. On pre-contrast T1-weighted black blood images (1G, long axis view), the mass (*) was relatively isointense but heterogeneous in signal intensity compared to adjacent normal myocardium. Following gadolinium-chelate contrast administration, the mass (*) enhanced heterogeneously (1H). RV = right ventricle; LV = left ventricle

Discussion

Cardiac myxomas are benign tumors that typically present during adulthood (within the third to sixth decades of life) and more commonly in women.[1-3] Most cases (90%) are sporadic, but some may be hereditary, notably as part of the Carney complex which is an autosomal dominant, multiple neoplasia syndrome.[4] Myxomas originate from the endocardium and typically project into the cardiac chamber. Most cardiac myxomas occur in the left atrium (59-75%), but may also be found in the right atrium (15-28%), right ventricle (3-8%) and left ventricle (0-4%).[1-3] Left atrial myxomas typically arise from the interatrial septa at the border of the fossa ovalis. Occasionally, they may also originate from the posterior wall of the left atrium or left atrial appendage. Rarely, cardiac myxoma may be multifocal (e.g. biatrial or biventricular).

As in this current case, cardiac myxomas are usually large at presentation, typically 5-6 cm in size.[1-3] They are usually round or oval shape with a lobular or smooth contour. In 16-34% of cases, however, the contour of the mass may be irregular and villous, with fragile gelatinous fronds that tend to fragment spontaneously. Most myxomas (85% [2]) are pedunculated and therefore mobile. These tumors are thought to arise from multipotential mesenchymal cells, which can differentiate into endothelial cells, smooth muscle, angioblasts, fibroblasts, cartilage, and myoblasts. There is suggestion that neuroendocrine differentiation may be responsible for non-cardiac related symptoms.[5] Approximately 10% of myxomas calcify. Cystic regions and hemorrhage may also be seen within cardiac myxomas.

Patients with cardiac myxomas often present with one or more features of the classic triad: intracardiac obstruction (67%); embolic events (29%); and constitutional or systemic symptoms (34%).[2] The likelihood for cardiac obstruction by a myxoma depends on its location, size and mobility.[1-3] If the tumor is mobile enough to cause acute obstruction of the atrioventricular valve or ventricular outflow tract, for example, syncope or even sudden death can occur. However, obstruction more commonly presents as mitral or tricuspid insufficiency, which may be temporary or positional from prolapse of the tumor. Permanent valvular damage may result from repetitive "wrecking ball" effect of the tumor on or across the valve. On physical exam, systolic and diastolic murmurs are auscultated in over 50% of patients.

Embolism is noted in 30-40% of patients with cardiac myxoma.[1-3] As most tumors reside within the left atrium, embolism is most frequently systemic—resulting in obstruction of the cerebral vessels, peripheral vessels, abdominal viscera, renal arteries, and/or coronary arteries. Right atrial tumors can cause pulmonary embolus, but these are rare. Embolisms may consist of portions of the myxoma itself or of thrombus, which often is seen in association with myxomas.

Constitutional symptoms in patients with cardiac myxoma include fatigue, fever, erythematous rash, arthralgias, myalgias, and weight loss. [1-3] These symptoms occur irrespective of tumor size or location. Laboratory abnormalities are anemia, increase in erythrocyte sedimentation rate, C-reactive protein, and globulin levels. Less common are increase in white blood cell count, thrombocytopenia, cyanosis, clubbing and Raynaud's.

In many patients, electrocardiograms are performed but are frequently nonspecific. As in this case of left atrial myxoma, chest radiography may show evidence of elevated left atrial pressure (53% [3]), which includes left atrial enlargement, pulmonary vascular redistribution, prominence of the pulmonary trunk, pulmonary edema and enlargement of the left atrial appendage. Pleural effusions and tumoral calcification may also be seen, albeit less commonly. However, chest radiographs are normal in over a third of patients.[3]

The proper identification of cardiac myxomas typically requires cross sectional imaging.[1,3,6,7] While echocardiography will often detect a cardiac mass, the performance of a CT and/or MR examination is often prudent as most cardiac masses are metastatic, requiring more extensive evaluation for lymphadenopathy and other visceral involvement. On CT, the size, location and morphology of a cardiac myxoma are typically well evaluated. The mass is typically isodense to myocardium but may be heterogeneous in its appearance if it contains calcified or cystic regions. The MRI appearance of cardiac myxomas is variable, most often somewhat heterogeneous on T1-weighted and T2-weighted images. Likewise, heterogeneous enhancement may be seen following Gd-chelate contrast administration. However, MRI is helpful for the evaluation of the tumor's mobility during the cardiac cycle. Cardiac-gated cine bright blood imaging (e.g. cine gradient echo and cine steady state free precession pulse sequences) provide cine viewing of the mechanical effects of the mass, of blood flow around the mass, of blood flow across the valves over the entire cardiac cycle (i.e. during systole and diastole).

Generally, MRI has better ability to characterize tissue than CT but this may not necessarily be helpful as many cardiac tumors have overlapping imaging features. Newer techniques using cardiac-gated multi-detector or electron beam CT may also provide similar cine information to that of cine bright blood MRI imaging, but MRI has the distinct advantages of multi-planar imaging (e.g. long- and short-axis views, Figures 1E and 1F) and not requiring the use of ionizing radiation. Conventional angiography is associated with a risk of embolization especially if intracavitary catheterization is performed and is therefore generally not performed. However, catheter angiography may be indicated if evaluation of the coronary vessels is required to determine the vascular supply of the tumor and/or exclude the co-existence of coronary artery disease.

The differential considerations for a cardiac myxoma are that of an intracavitary cardiac mass, notably thrombus or a cardiac metastasis (e.g. renal cell carcinoma). Bland thrombus typically will not enhance following contrast administration, whether on CT or MRI. Cardiac thrombus is also more likely to occur in patients with underlying cardiac disease such as atrial fibrillation or a prior myocardial infarct. Thrombus is usually sessile and adherent to the myocardial wall and not likely to be mobile or pedunculated. Metastases often have

other features of metastatic disease such as mediastinal adenopathy and/or pulmonary metastases.

The diagnosis of primary cardiac tumor is typically made only after the potential of metastatic disease is excluded. Metastatic lesions to the heart are much more common than primary lesions. However, of the primary cardiac tumors, myxomas are the most common. Other intracavitary primary cardiac masses include sarcomas (second most common primary cardiac neoplasm) and papillary fibroelastomas and, but these are less common. Sarcomas often invade adjacent structures. Papillary fibroelastomas typically occur on the mitral or aortic valve and tend to be smaller.

The finding of a large enhancing left atrial mass in an adult woman without signs or evidence for extra-cardiac malignancy should suggest the diagnosis of atrial myxoma. The treatment is surgical excision. Due to the risk of sudden death from acute obstruction, these tumors should be excised without delay.

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